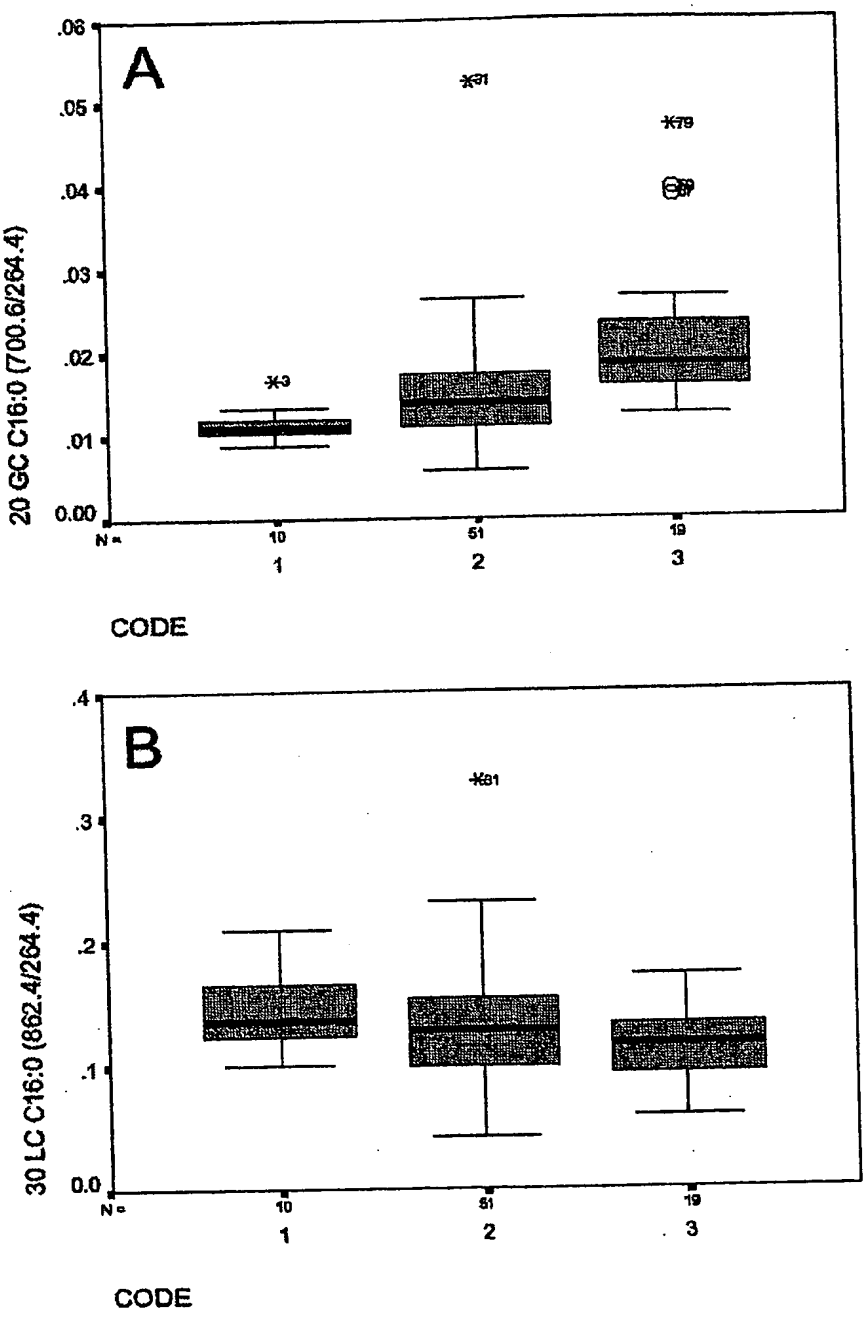
A
20 GC C16:0 (700.6/264.4)
.06
.05
.04
.03
.02
.01
0.00
N =
10
51
19
1
2
3
CODE
B
30 LC C16:0 (862.4/264.4)
.4
.3
.2
.1
0.0
N =
10
51
19
1
2
3
CODE

FIGURE 1

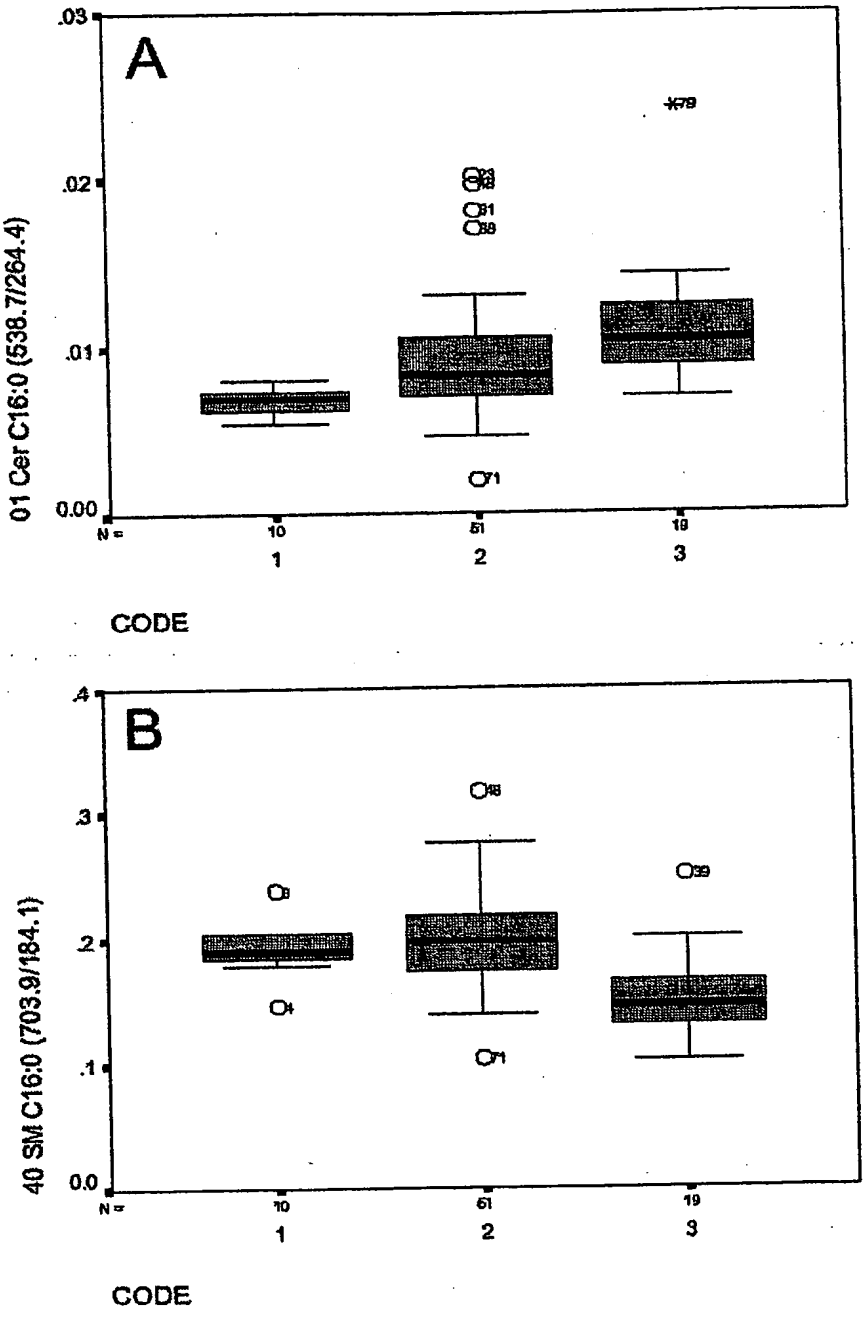
A
01 Cer C16:0 (538.7/264.4)
.03
.02
.01
0.00
N =
10
51
19
1
2
3
CODE
B
40 SM C16:0 (703.9/184.1)
.4
.3
.2
.1
0.0
N =
10
51
19
1
2
3
CODE

FIGURE 2

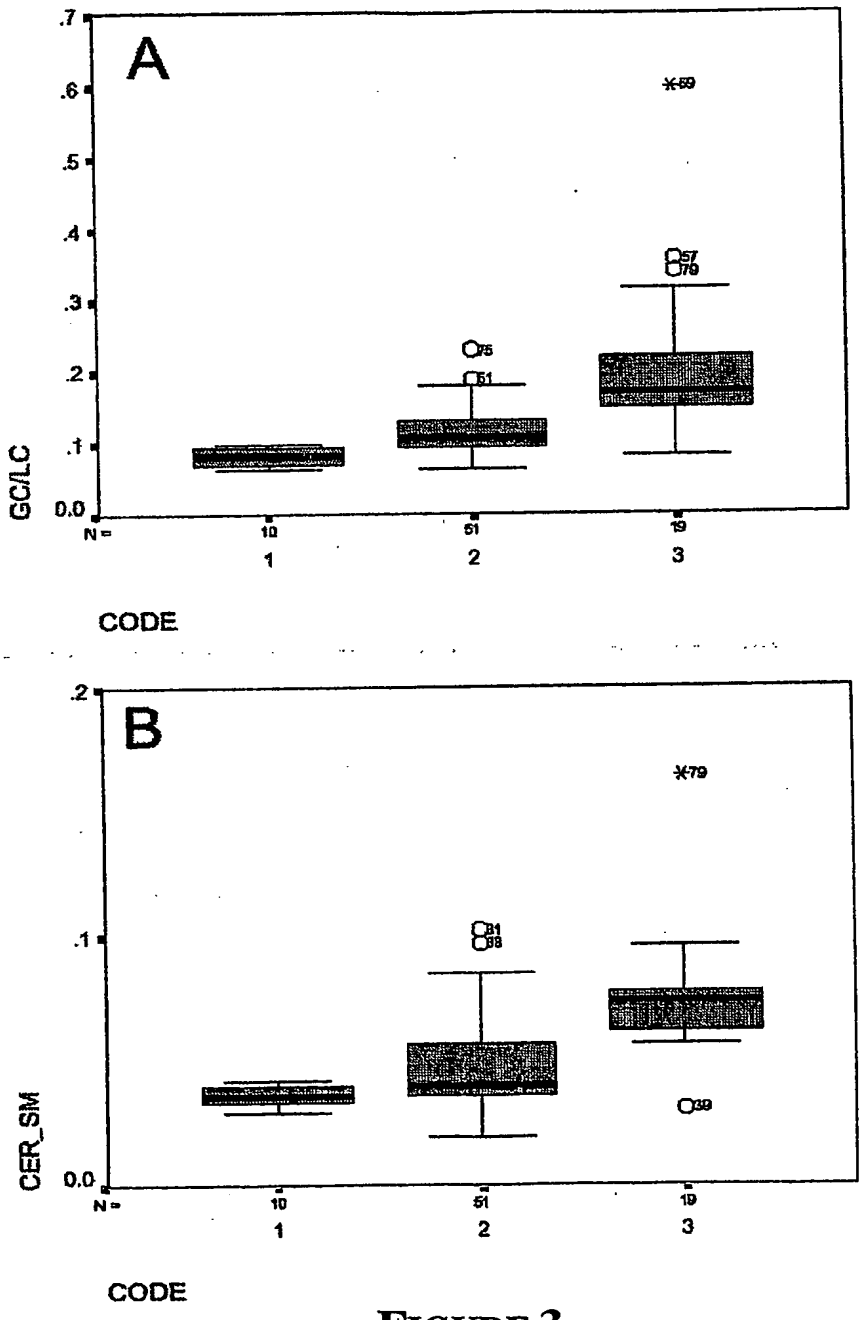
A
GC/LC
.7
.6
.5
.4
.3
.2
.1
0.0
N =
10
51
19
1
2
3
CODE
B
CER_SM
.2
.1
0.0
N =
10
51
19
1
2
3
CODE

FIGURE 3

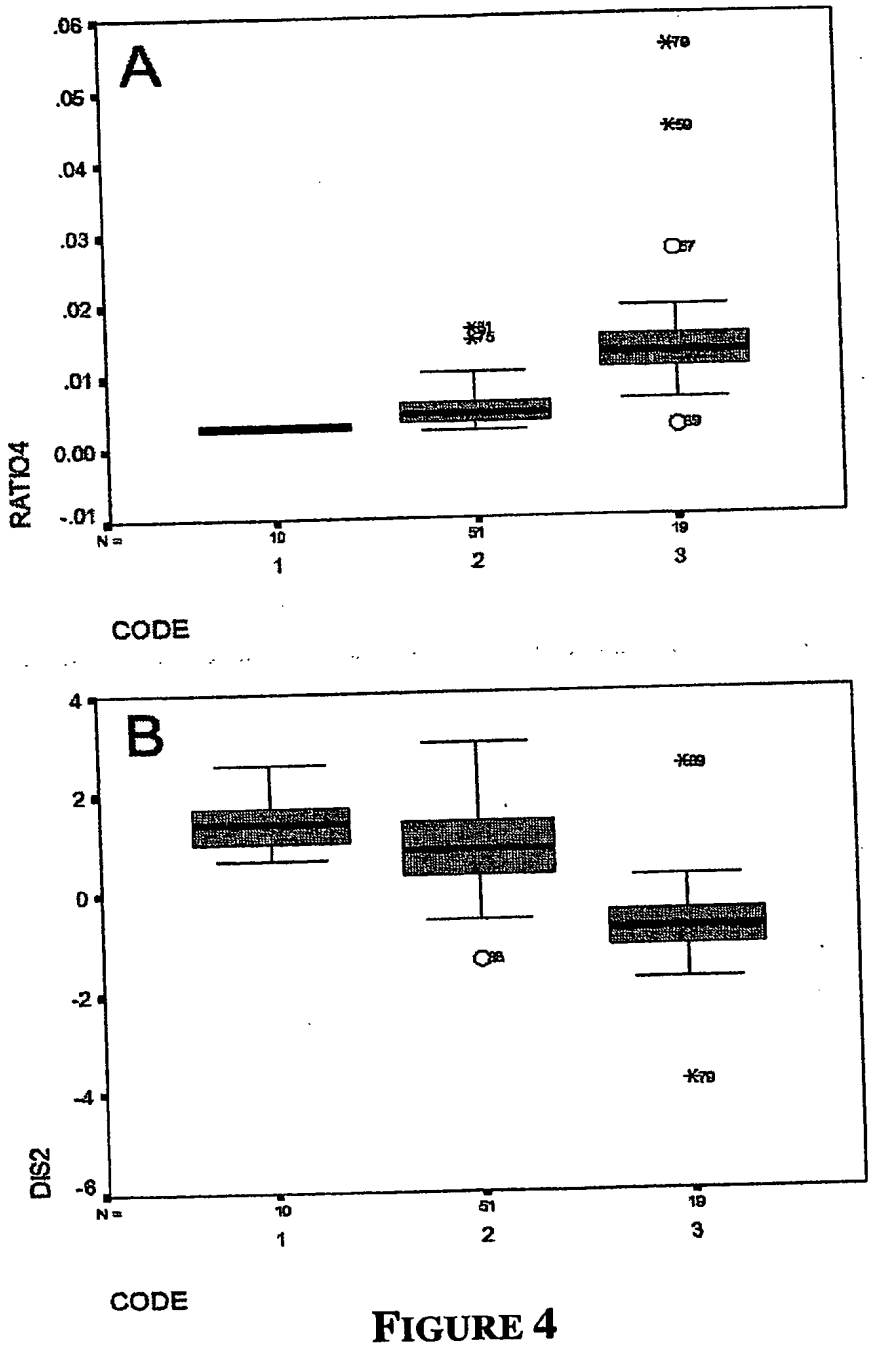
A
.06
.05
.04
.03
.02
.01
0.00
-.01
RATIO4
N =
10
51
19
1
2
3
CODE
B
4
2
0
-2
-4
-6
DIS2
N =
10
51
19
1
2
3
CODE

FIGURE 4

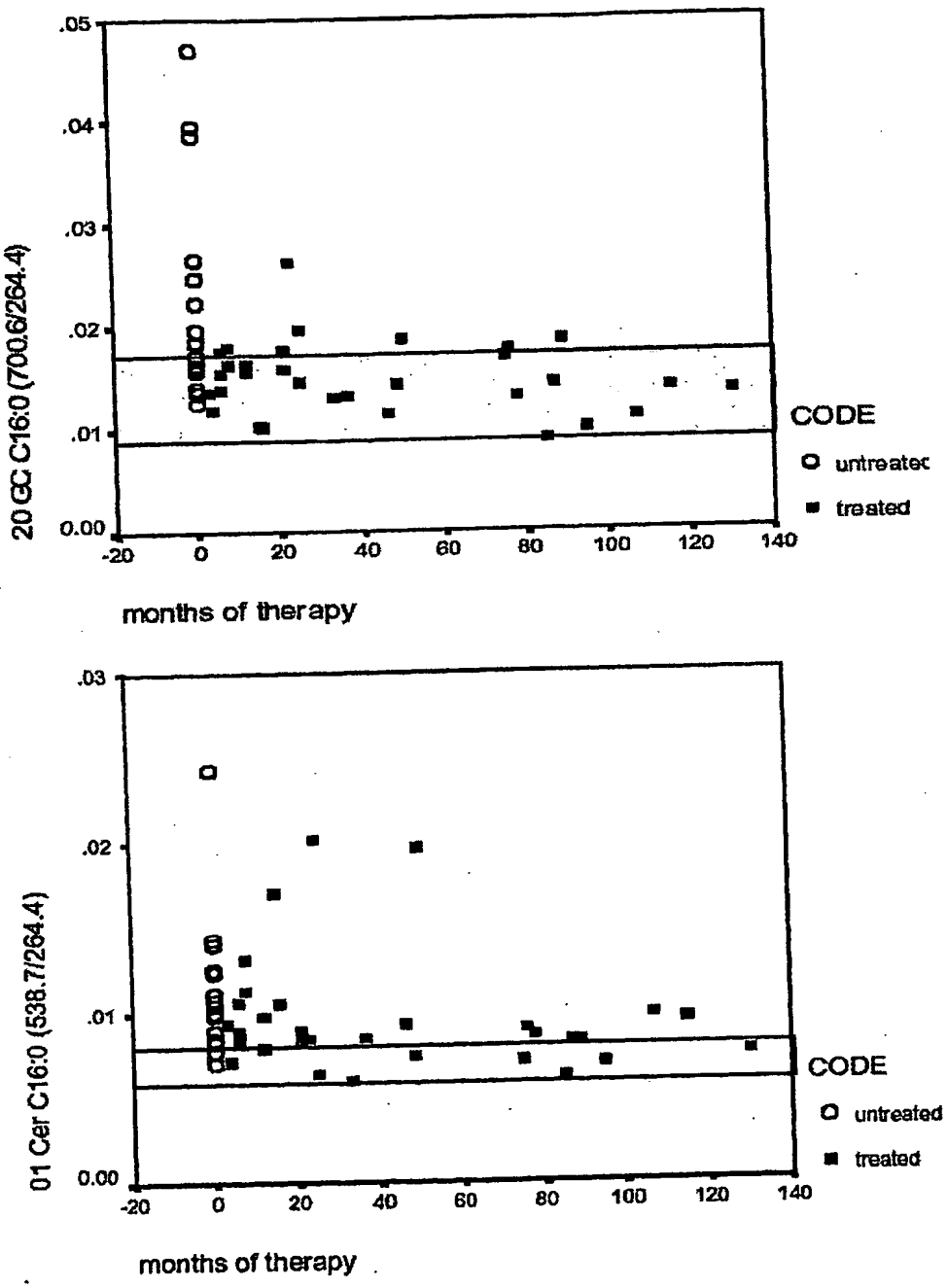
20 GC C16:0 (700.6/264.4)
.05
.04
.03
.02
.01
0.00
-20
0
20
40
60
80
100
120
140
months of therapy
CODE
untreated
treated
01 Cer C16:0 (538.7/264.4)
.03
.02
.01
0.00
-20
0
20
40
60
80
100
120
140
months of therapy
CODE
untreated
treated

FIGURE 5

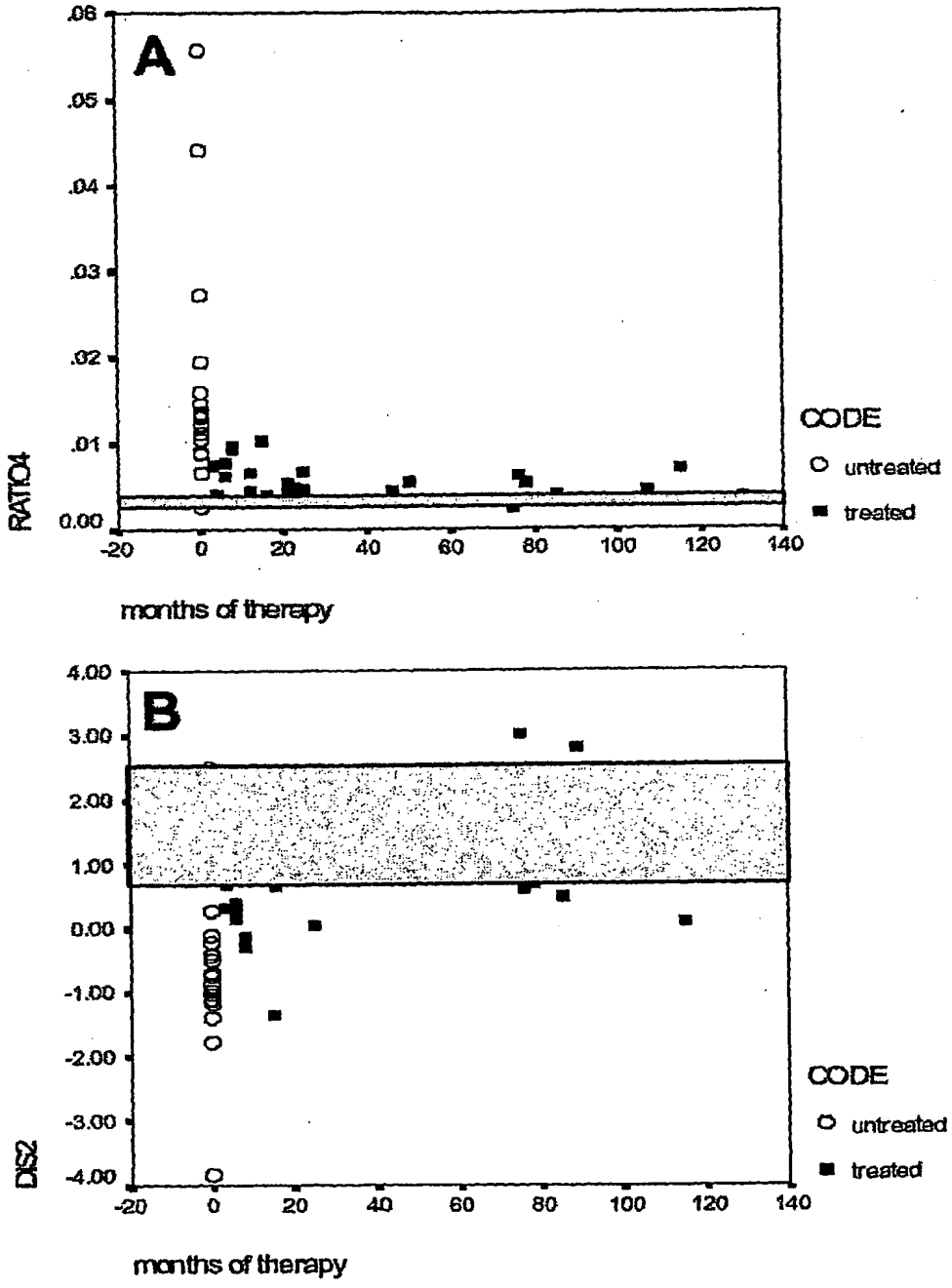
A
.06
.05
.04
.03
.02
.01
0.00
RATIO4
-20
0
20
40
60
80
100
120
140
months of therapy
CODE
untreated
treated
B
4.00
3.00
2.00
1.00
0.00
-1.00
-2.00
-3.00
-4.00
DIS2
-20
0
20
40
60
80
100
120
140
months of therapy
CODE
untreated
treated

FIGURE 6

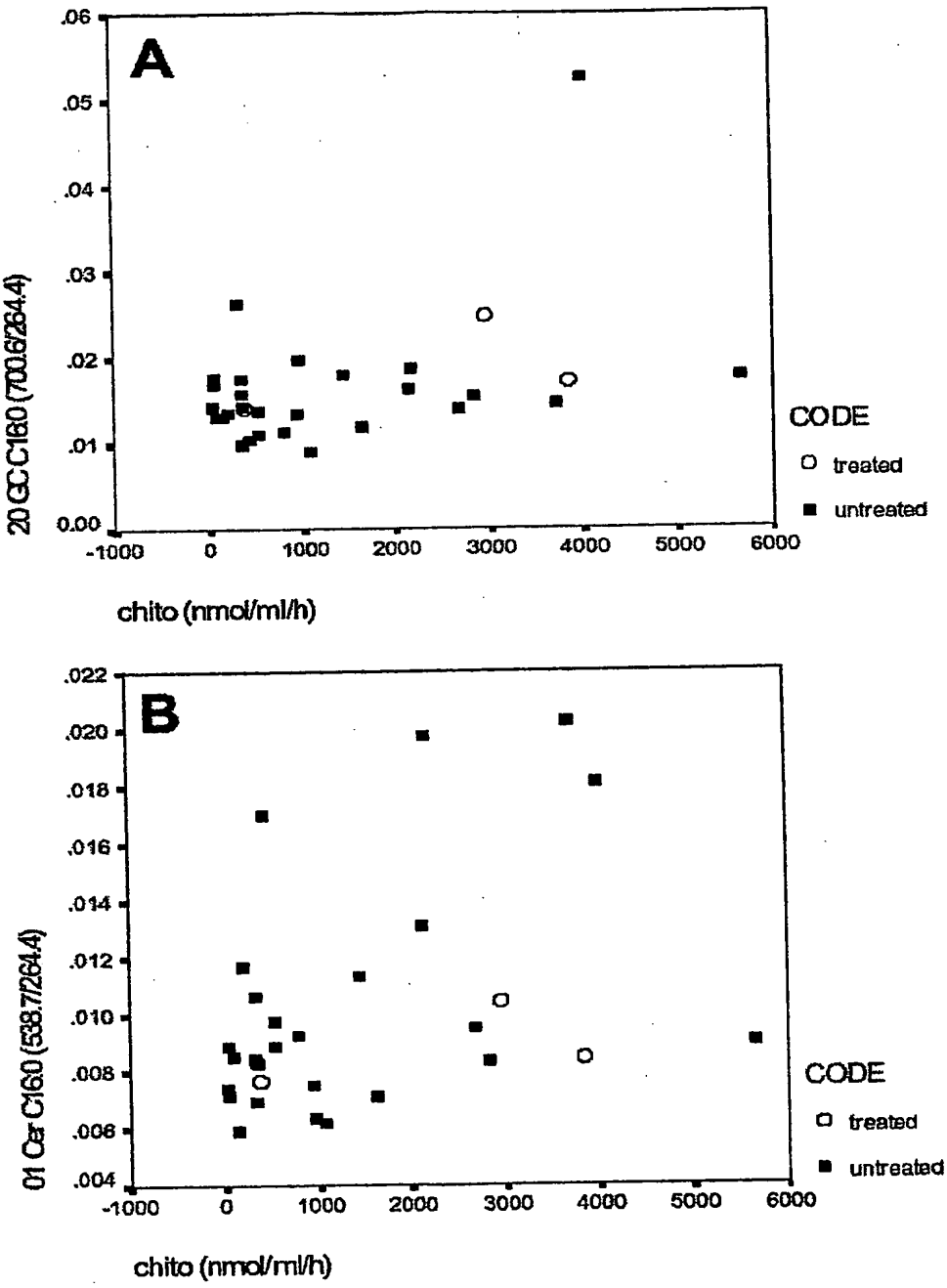
A
.06
.05
.04
.03
.02
.01
0.00
20 GCC C16:0 (700.6/264.4)
-1000
0
1000
2000
3000
4000
5000
6000
chito (nmol/ml/h)
CODE
treated
untreated
B
.022
.020
.018
.016
.014
.012
.010
.008
.006
.004
01 Cer C16:0 (538.7/264.4)
-1000
0
1000
2000
3000
4000
5000
6000
chito (nmol/ml/h)
CODE
treated
untreated

FIGURE 7

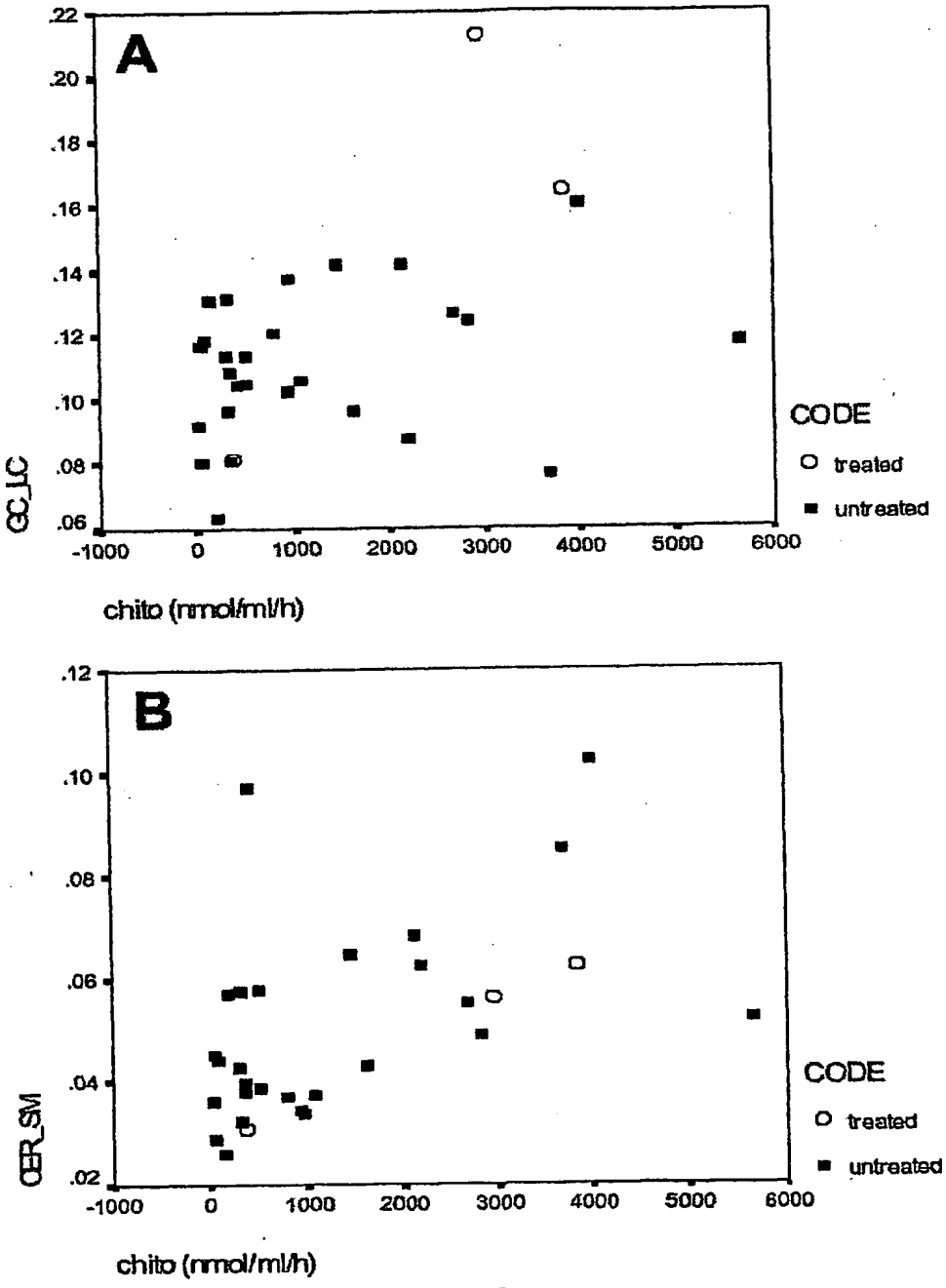
A
.22
.20
.18
.16
.14
.12
.10
.08
.06
QC_LC
-1000
0
1000
2000
3000
4000
5000
6000
chito (nmol/ml/h)
CODE
treated
untreated
B
.12
.10
.08
.06
.04
.02
CER_SM
-1000
0
1000
2000
3000
4000
5000
6000
chito (nmol/ml/h)
CODE
treated
untreated

FIGURE 8

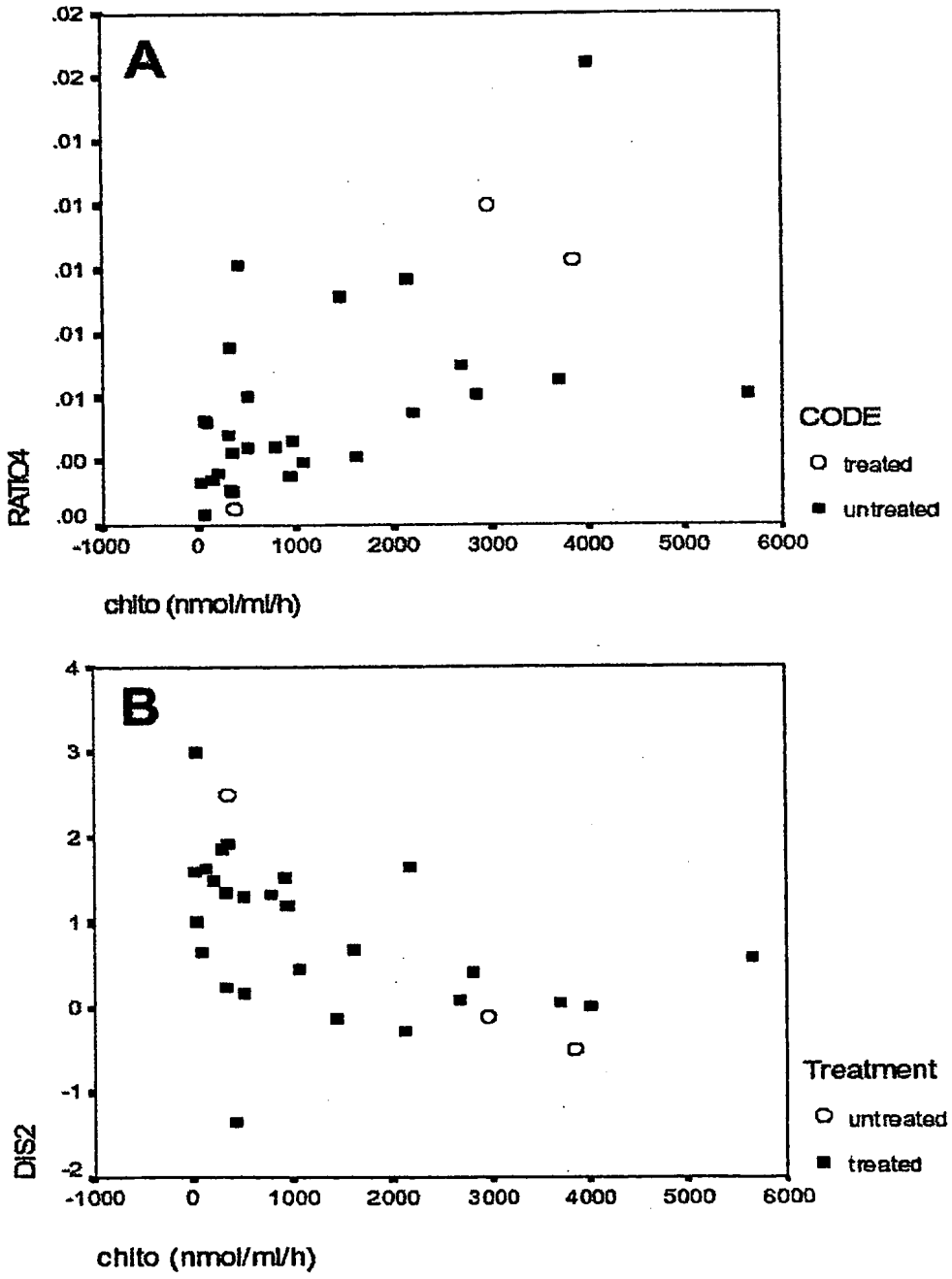
A
RATIO4
.02
.02
.01
.01
.01
.01
.01
.00
.00
-1000
0
1000
2000
3000
4000
5000
6000
chito (nmol/ml/h)
CODE
treated
untreated
B
DIS2
4
3
2
1
0
-1
-2
-1000
0
1000
2000
3000
4000
5000
6000
chito (nmol/ml/h)
Treatment
untreated
treated

FIGURE 9

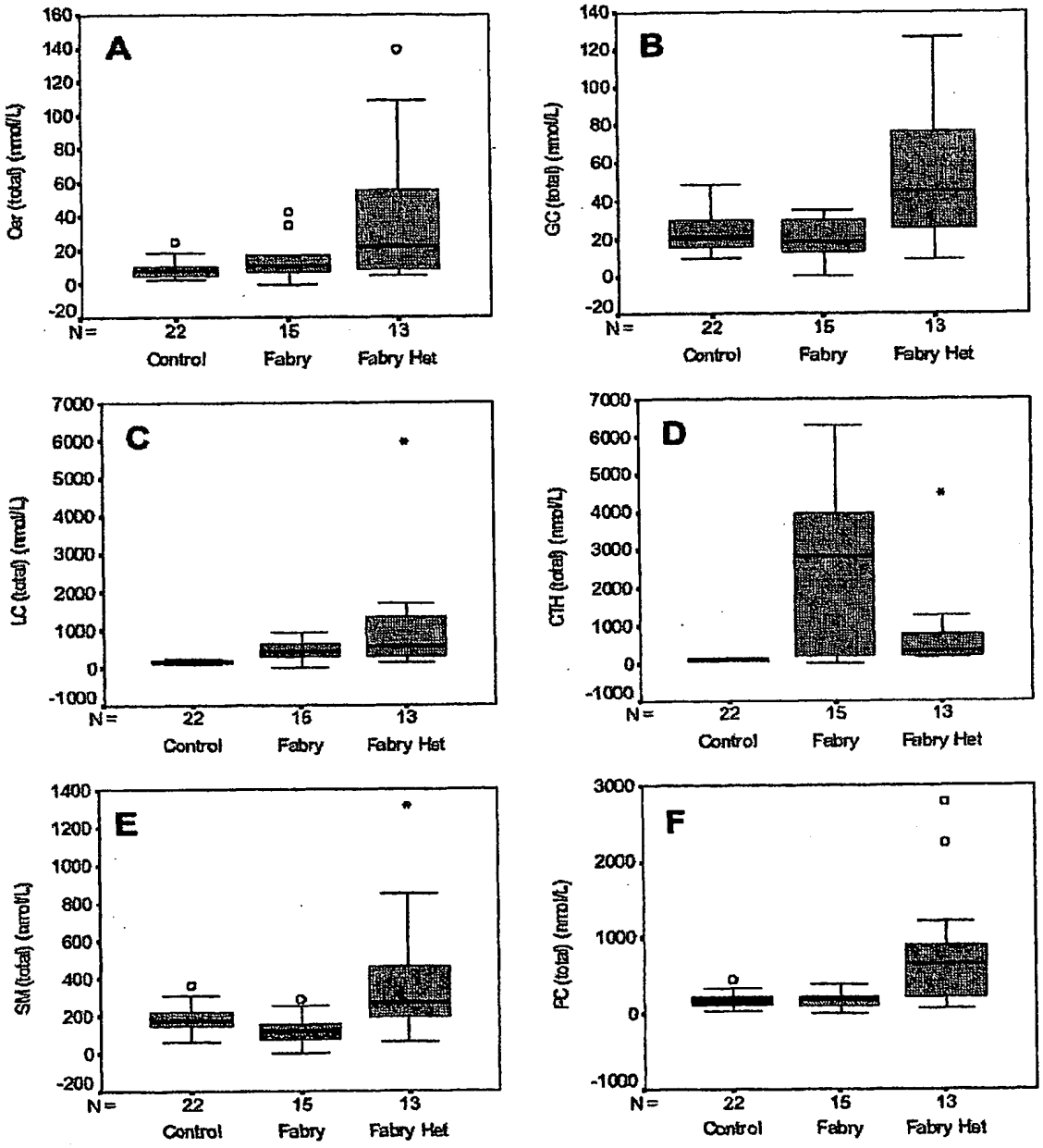
A
Cer (total) (nmol/L)
160
140
120
100
80
60
40
20
0
-20
N= 22 15 13
Control Fabry Fabry Het
B
GC (total) (nmol/L)
140
120
100
80
60
40
20
0
-20
N= 22 15 13
Control Fabry Fabry Het
C
LC (total) (nmol/L)
7000
6000
5000
4000
3000
2000
1000
0
-1000
N= 22 15 13
Control Fabry Fabry Het
D
CTH (total) (nmol/L)
7000
6000
5000
4000
3000
2000
1000
0
-1000
N= 22 15 13
Control Fabry Fabry Het
E
SM (total) (nmol/L)
1400
1200
1000
800
600
400
200
0
-200
N= 22 15 13
Control Fabry Fabry Het
F
PC (total) (nmol/L)
3000
2000
1000
0
-1000
N= 22 15 13
Control Fabry Fabry Het

FIGURE 10

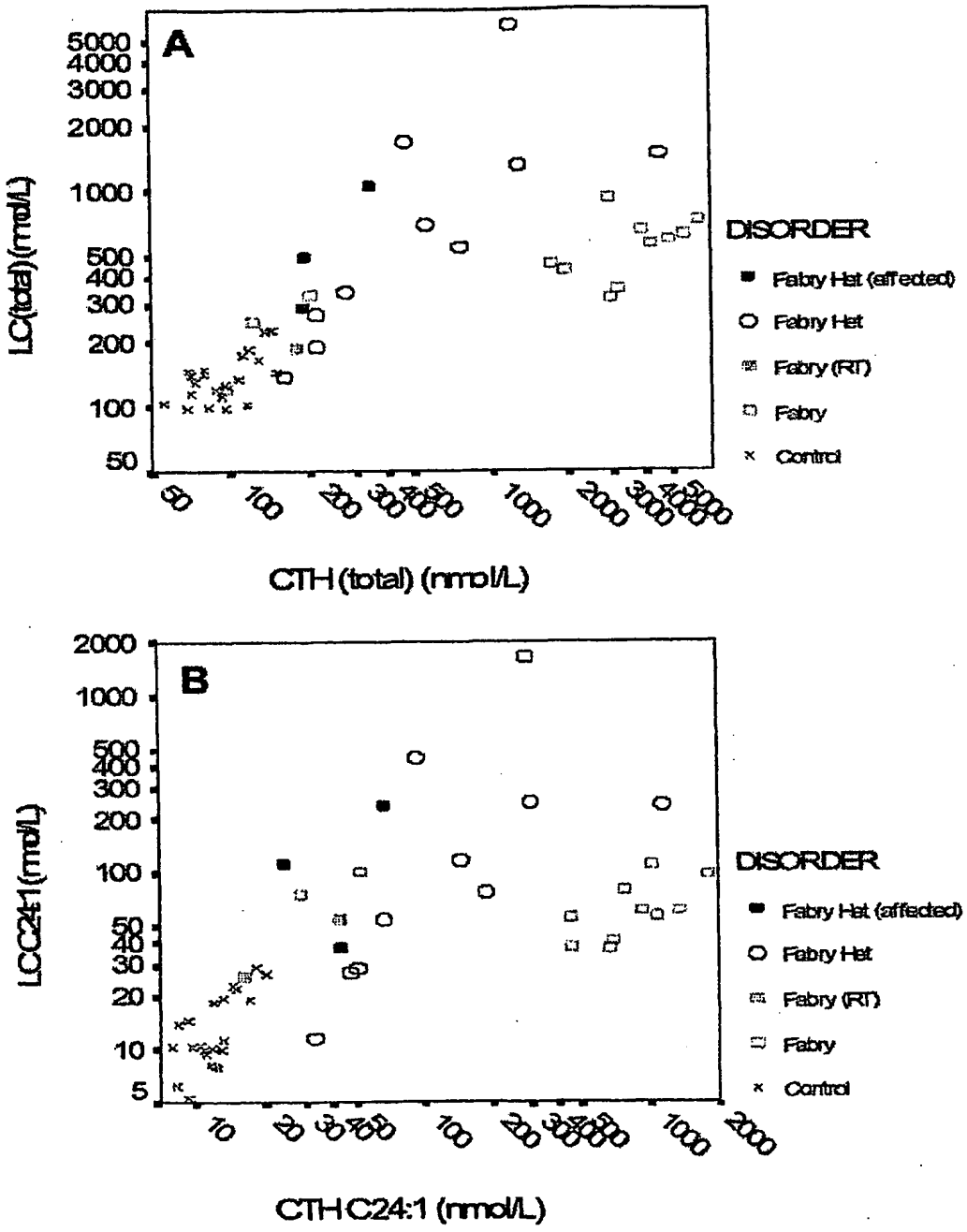
A
5000
4000
3000
2000
1000
500
400
300
200
100
50
LC(total)(nmol/L)
50
100
200
300
400
500
1000
2000
3000
4000
5000
CTH (total) (nmol/L)
DISORDER
Fabry Het (affected)
Fabry Het
Fabry (RT)
Fabry
Control
B
2000
1000
500
400
300
200
100
50
40
30
20
10
5
LCC24:1(nmol/L)
10
20
30
40
50
100
200
300
400
500
1000
2000
CTH C24:1 (nmol/L)
DISORDER
Fabry Het (affected)
Fabry Het
Fabry (RT)
Fabry
Control

FIGURE 11

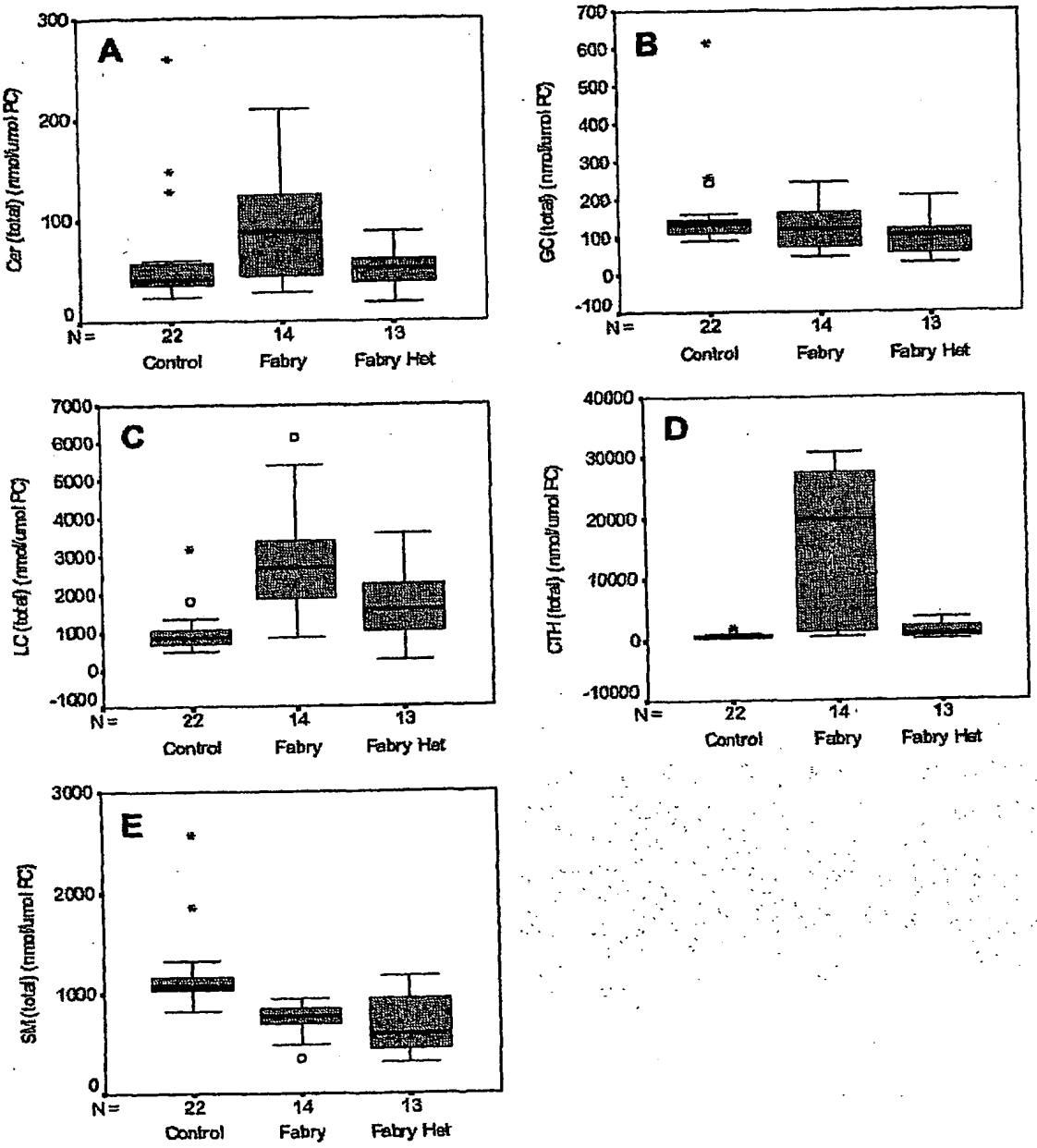
A
Cer (total) (nmol/umol PC)
300
200
100
0
N=
22
14
13
Control
Fabry
Fabry Het
B
GC (total) (nmol/umol PC)
700
600
500
400
300
200
100
0
-100
C
LC (total) (nmol/umol PC)
7000
6000
5000
4000
3000
2000
1000
0
-1000
D
CTH (total) (nmol/umol PC)
40000
30000
20000
10000
0
-10000
E
SM (total) (nmol/umol PC)
3000
2000
1000
0

FIGURE 12

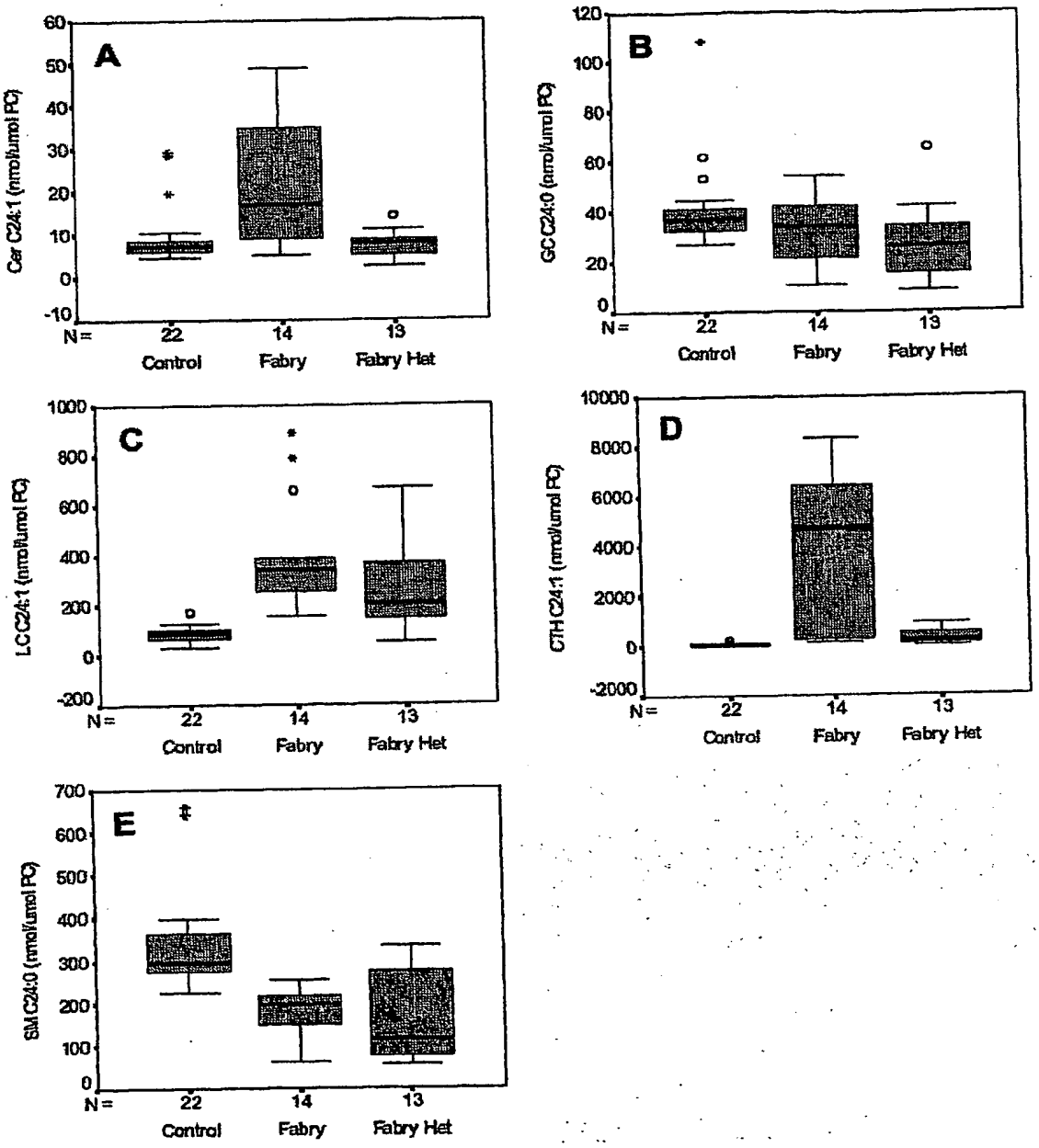
A
Cer C24:1 (nmol/umol PC)
60
50
40
30
20
10
0
-10
N=
22
14
13
Control
Fabry
Fabry Het
B
GCC24:0 (nmol/umol PC)
120
100
80
60
40
20
0
N=
22
14
13
Control
Fabry
Fabry Het
C
LCC24:1 (nmol/umol PC)
1000
800
600
400
200
0
-200
N=
22
14
13
Control
Fabry
Fabry Het
D
CTH C24:1 (nmol/umol PC)
10000
8000
6000
4000
2000
0
-2000
N=
22
14
13
Control
Fabry
Fabry Het
E
SM C24:0 (nmol/umol PC)
700
600
500
400
300
200
100
0
N=
22
14
13
Control
Fabry
Fabry Het

FIGURE 13

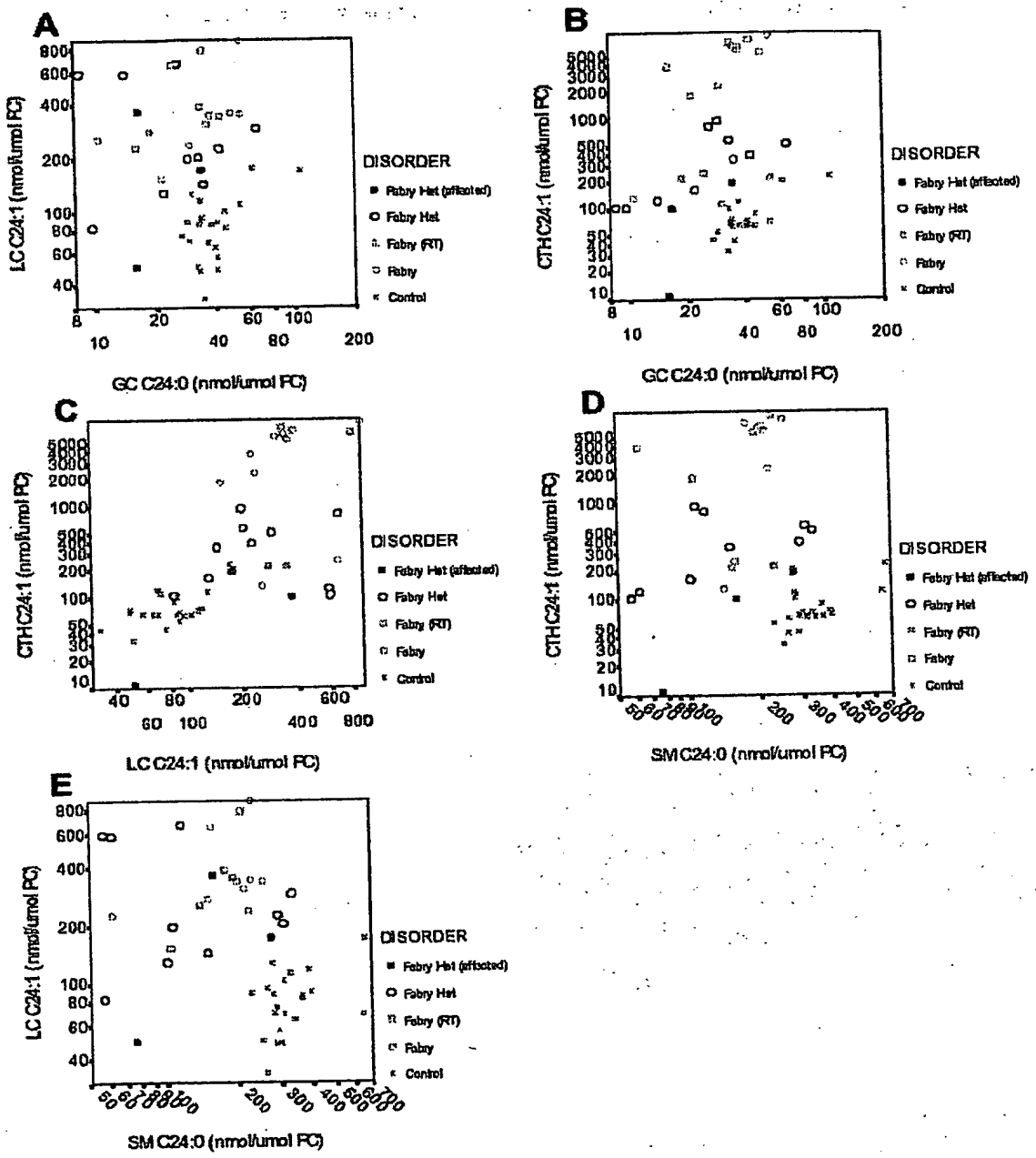
A
LC C24:1 (nmol/umol PC)
GC C24:0 (nmol/umol PC)
B
CTHC24:1 (nmol/umol PC)
GC C24:0 (nmol/umol PC)
C
CTHC24:1 (nmol/umol PC)
LC C24:1 (nmol/umol PC)
D
CTHC24:1 (nmol/umol PC)
SM C24:0 (nmol/umol PC)
E
LC C24:1 (nmol/umol PC)
SM C24:0 (nmol/umol PC)
DISORDER
Fabry Het (affected)
Fabry Het
Fabry (RT)
Fabry
Control

FIGURE 14

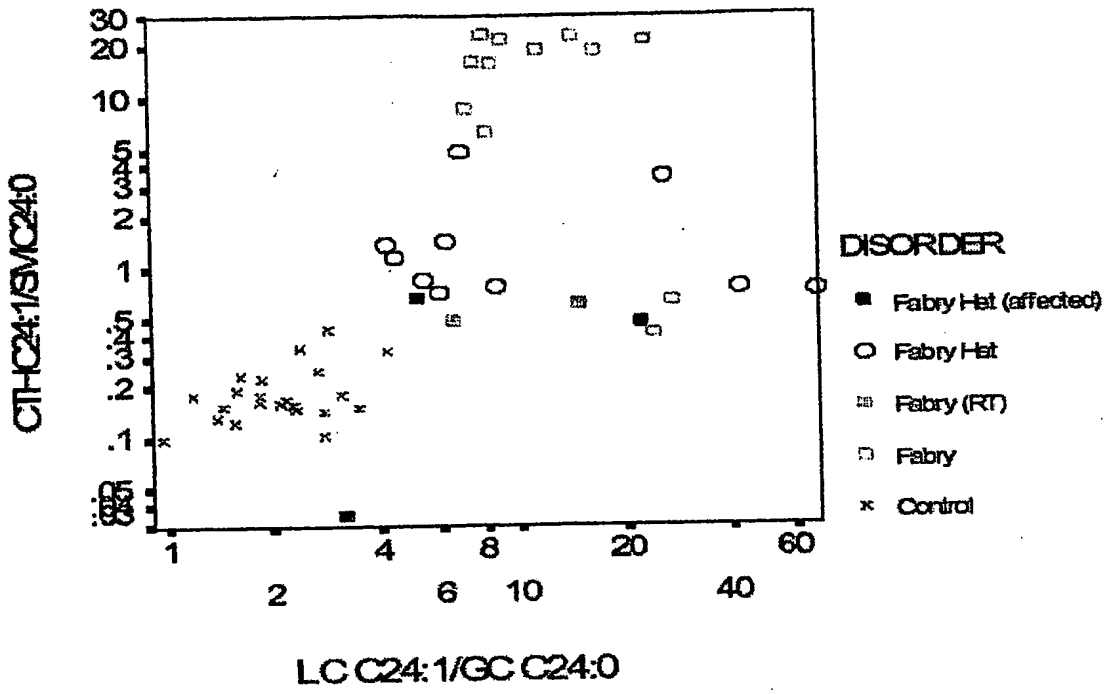
CTHC24:1/SMC24:0
LC C24:1/GC C24:0
DISORDER
Fabry Het (affected)
Fabry Het
Fabry (RT)
Fabry
Control

FIGURE 15

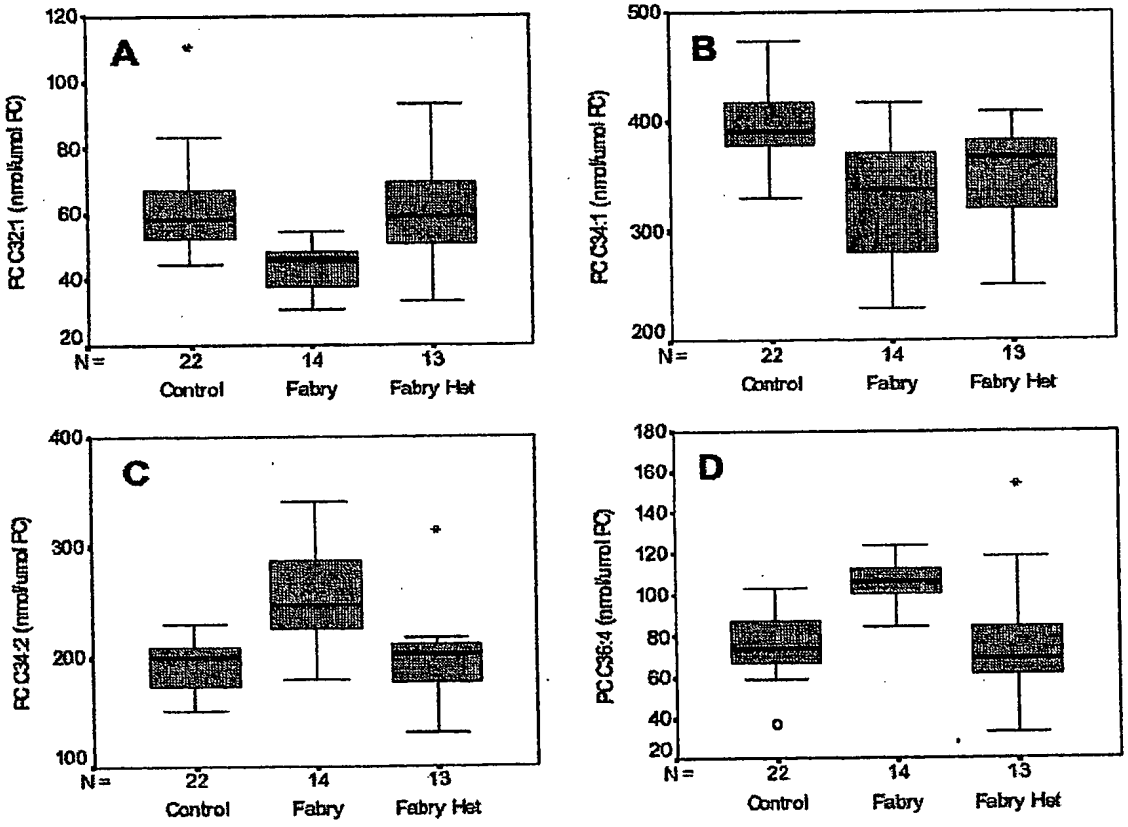
A
PC C32:1 (nmol/umol PC)
120
100
80
60
40
20
N=
22
14
13
Control
Fabry
Fabry Het
B
PC C34:1 (nmol/umol PC)
500
400
300
200
C
PC C34:2 (nmol/umol PC)
400
300
200
100
D
PC C36:4 (nmol/umol PC)
180
160
140
120
100
80
60
40
20

FIGURE 16

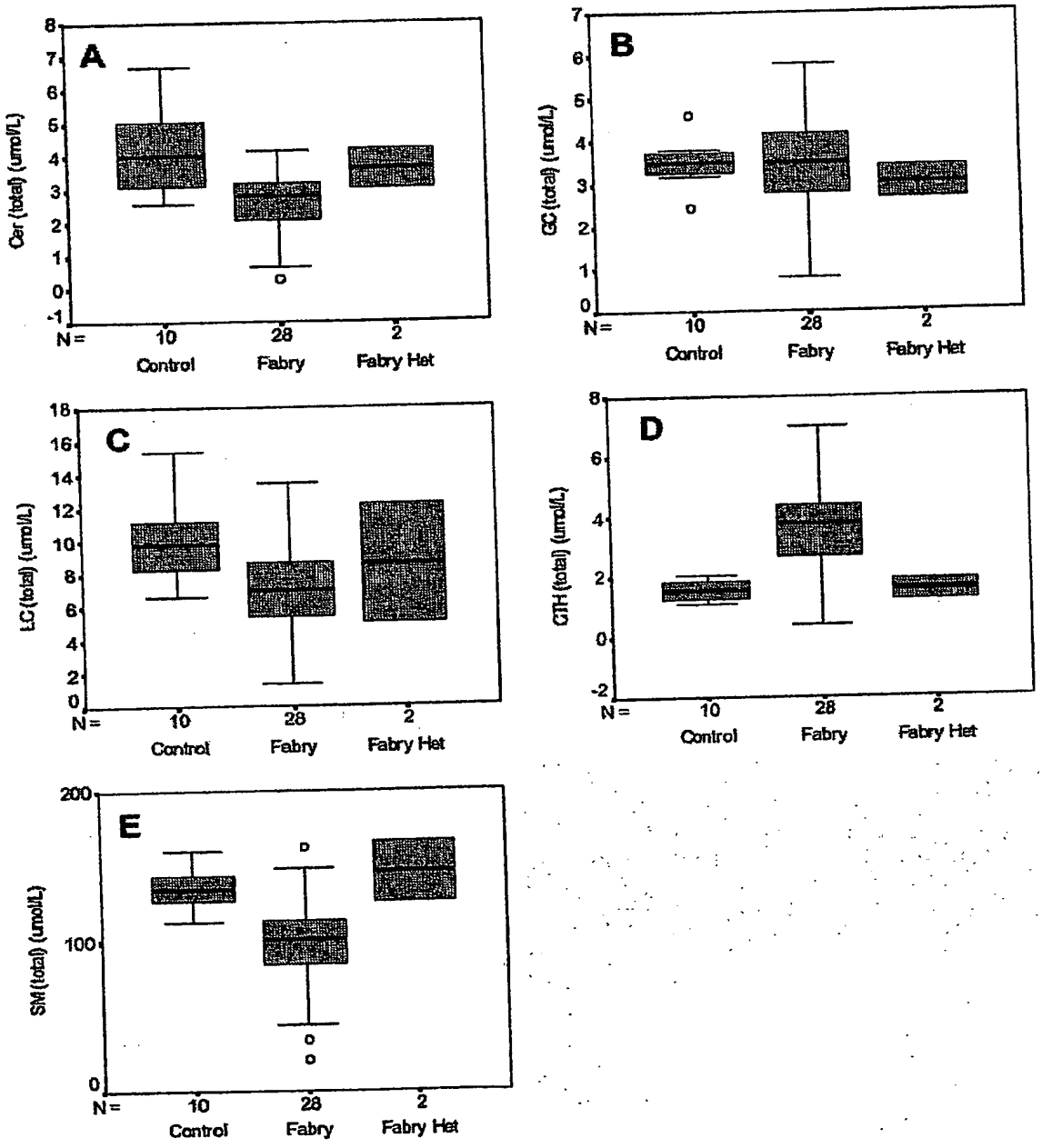
A
Cer (total) (umol/L)
N=
10
28
2
Control
Fabry
Fabry Het
B
GC (total) (umol/L)
C
LC (total) (umol/L)
D
GTH (total) (umol/L)
E
SM (total) (umol/L)

FIGURE 17

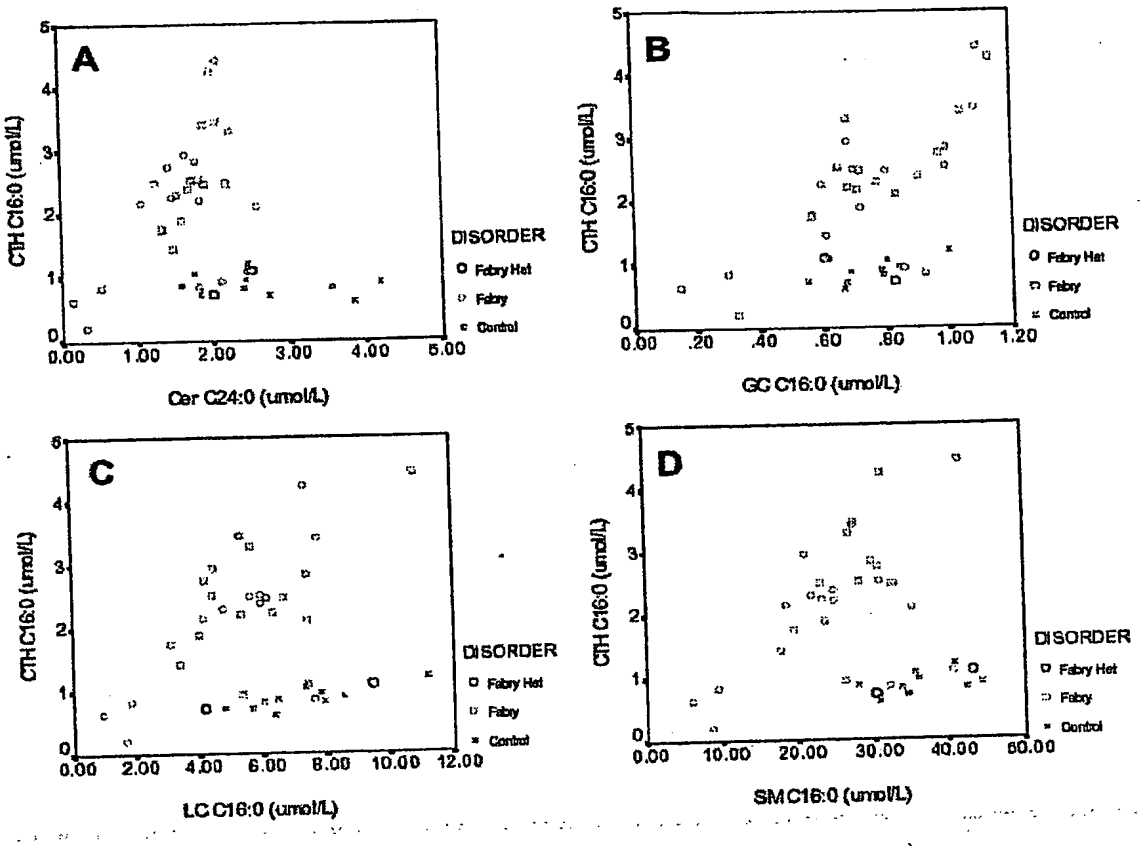
A
CTH C16:0 (umol/L)
Cer C24:0 (umol/L)
DISORDER
Fabry Het
Fabry
Control
B
CTH C16:0 (umol/L)
GC C16:0 (umol/L)
DISORDER
Fabry Het
Fabry
Control
C
CTH C16:0 (umol/L)
LC C16:0 (umol/L)
DISORDER
Fabry Het
Fabry
Control
D
CTH C16:0 (umol/L)
SM C16:0 (umol/L)
DISORDER
Fabry Het
Fabry
Control

FIGURE 18

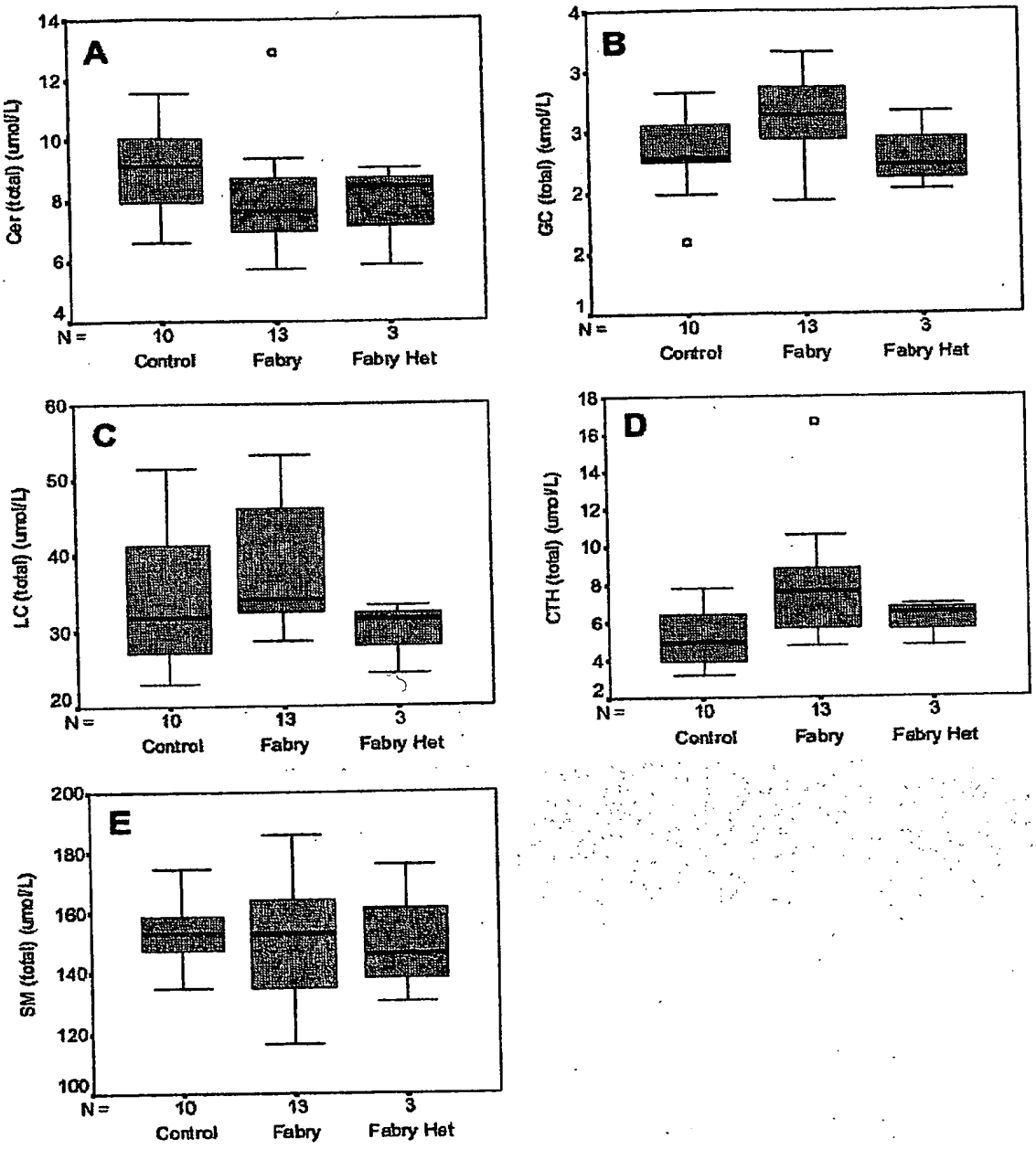
A
Cer (total) (umol/L)
14
12
10
8
6
4
N =
10
13
3
Control
Fabry
Fabry Het
B
GC (total) (umol/L)
4
3
3
2
2
1
N =
10
13
3
Control
Fabry
Fabry Het
C
LC (total) (umol/L)
60
50
40
30
20
N =
10
13
3
Control
Fabry
Fabry Het
D
CTH (total) (umol/L)
18
16
14
12
10
8
6
4
2
N =
10
13
3
Control
Fabry
Fabry Het
E
SM (total) (umol/L)
200
180
160
140
120
100
N =
10
13
3
Control
Fabry
Fabry Het

FIGURE 19

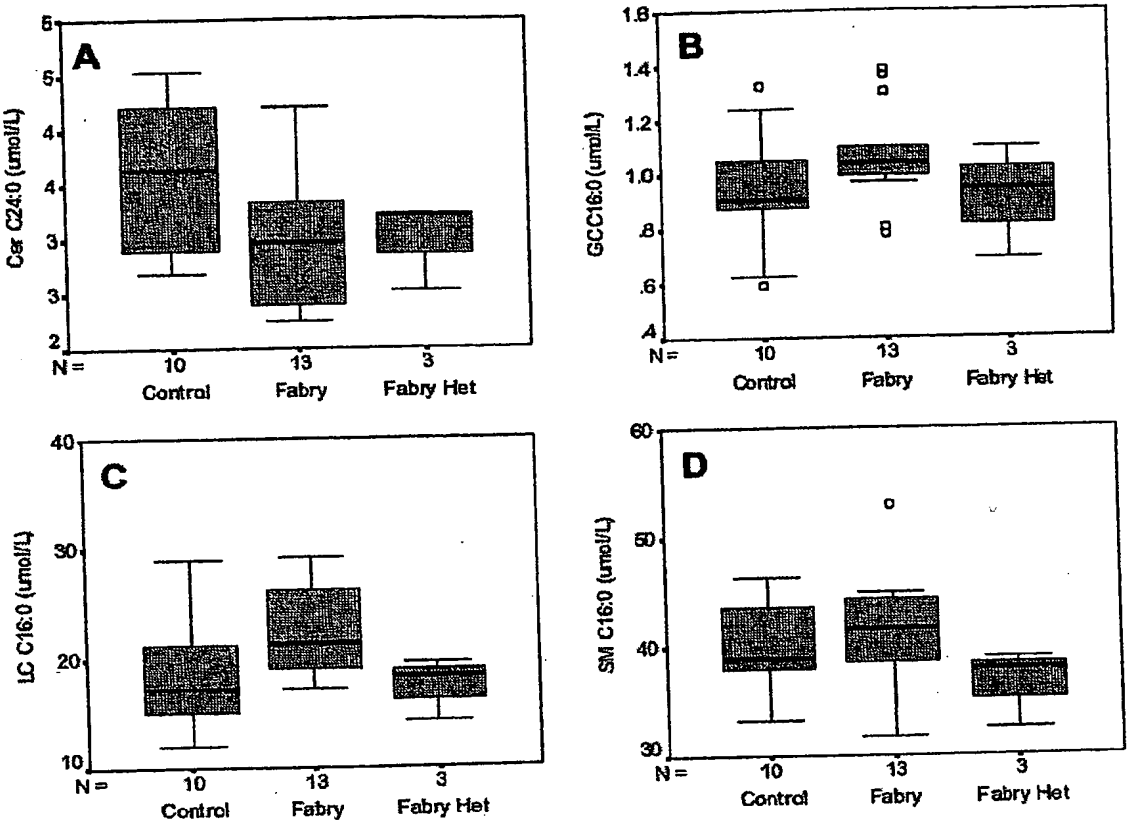
A
Car C24:0 (umol/L)
N =
10
13
3
Control
Fabry
Fabry Het
B
GCC16:0 (umol/L)
C
LC C16:0 (umol/L)
D
SM C16:0 (umol/L)

FIGURE 20

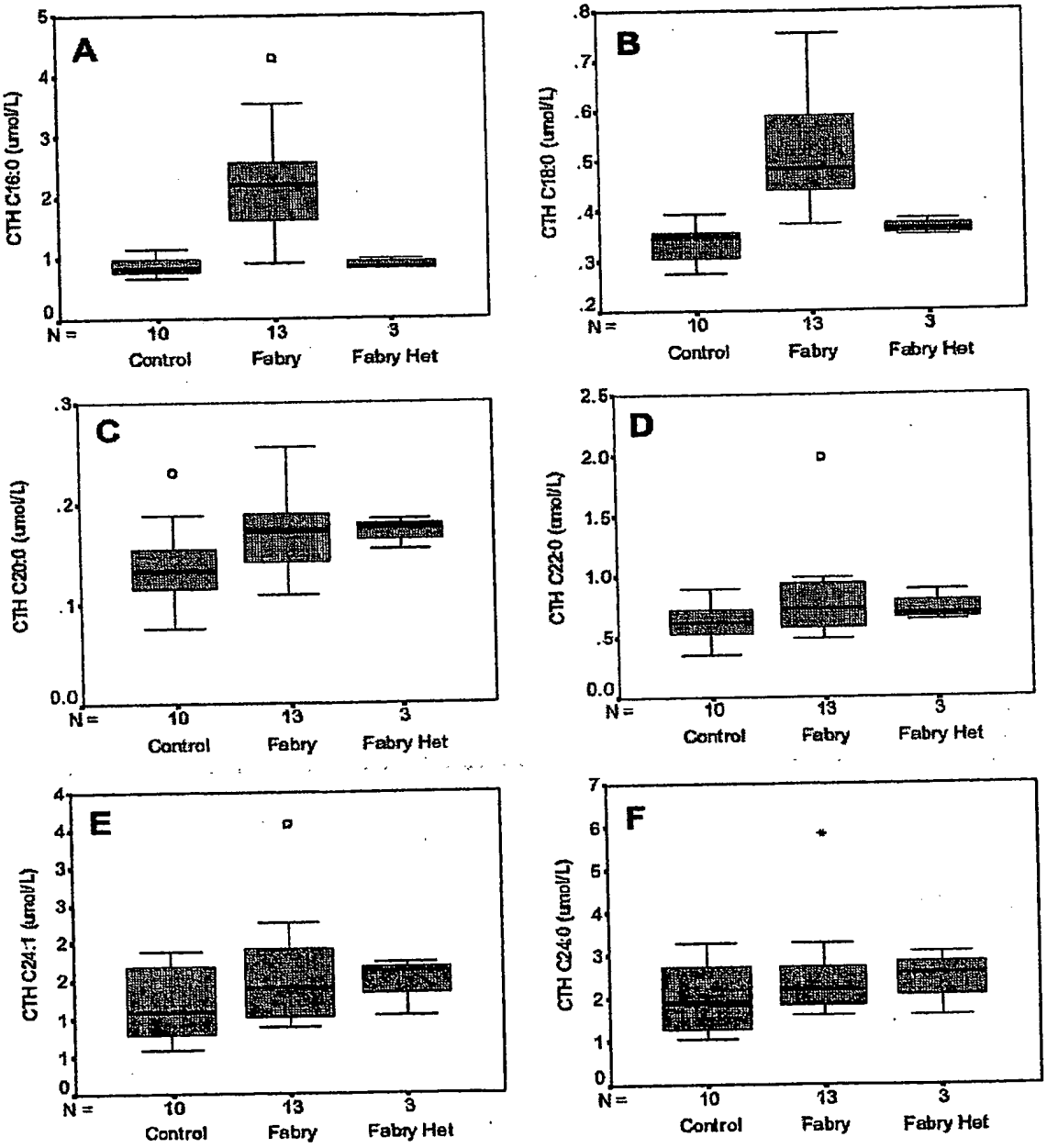
A
CTH C16:0 (umol/L)
N =
10
13
3
Control
Fabry
Fabry Het
B
CTH C18:0 (umol/L)
C
CTH C20:0 (umol/L)
D
CTH C22:0 (umol/L)
E
CTH C24:1 (umol/L)
F
CTH C24:0 (umol/L)

FIGURE 21

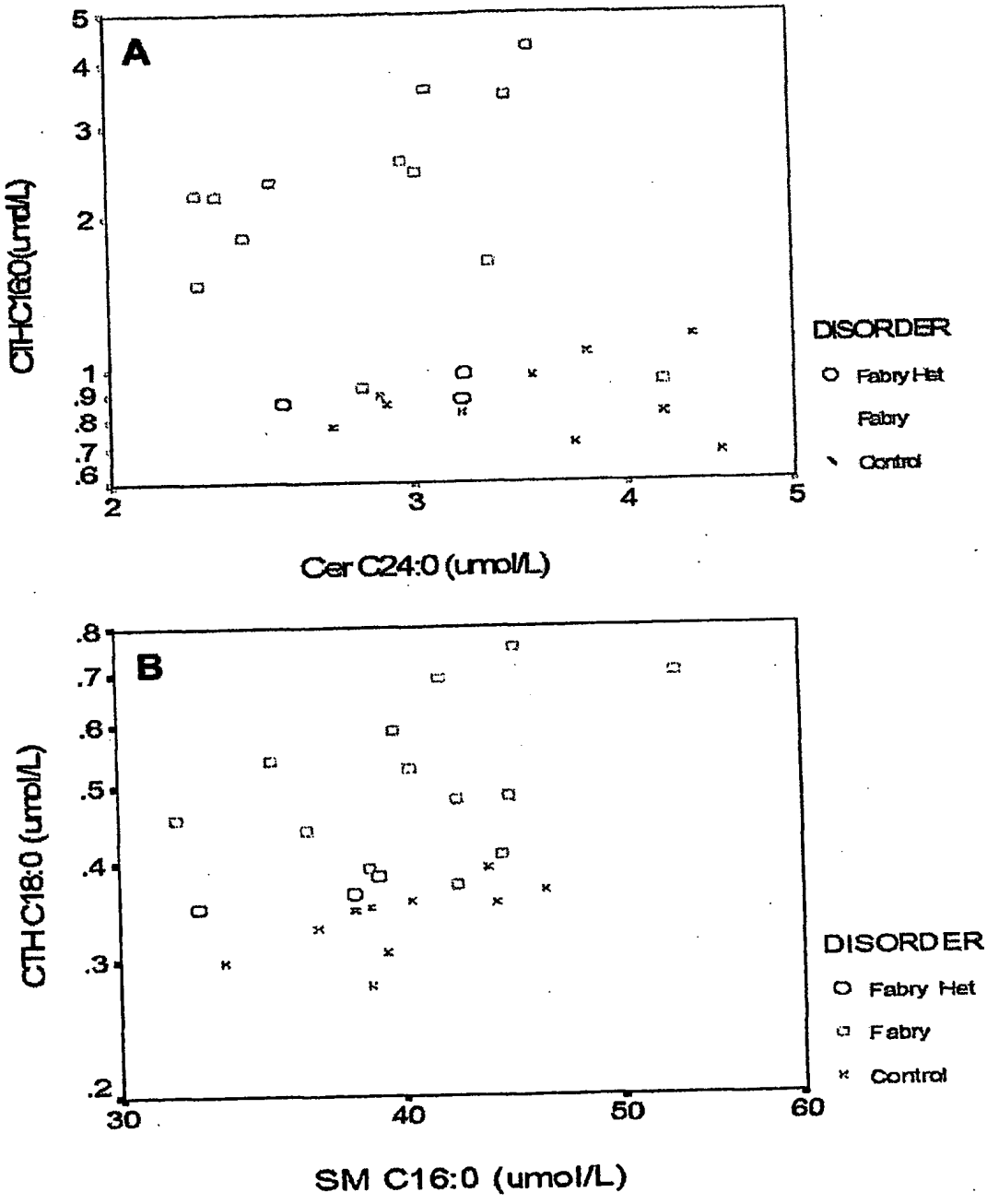
A
CTHC16O(umol/L)
Cer C24:0 (umol/L)
DISORDER
Fabry Het
Fabry
Control
B
CTH C18:0 (umol/L)
SM C16:0 (umol/L)
DISORDER
Fabry Het
Fabry
Control

FIGURE 22

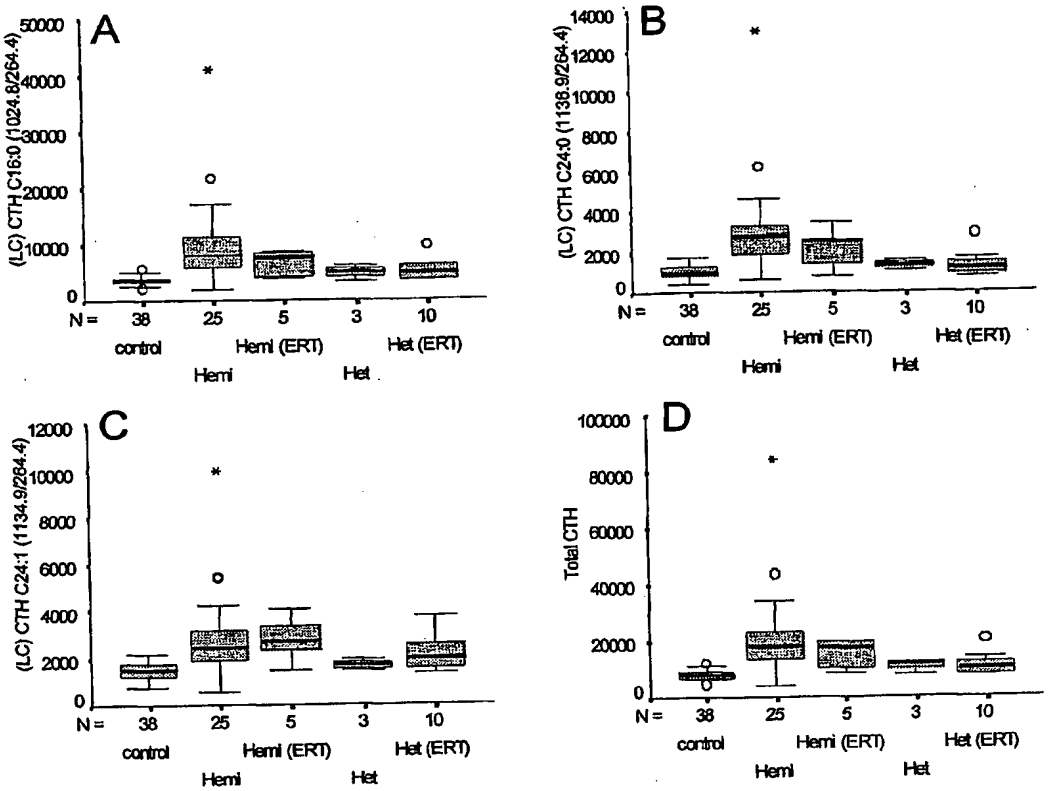
A
(LC) CTH C16:0 (1024.8/264.4)
50000
40000
30000
20000
10000
0
N = 38 25 5 3 10
control
Hemi (ERT)
Het (ERT)
Hemi
Het
B
(LC) CTH C24:0 (1138.9/264.4)
14000
12000
10000
8000
6000
4000
2000
0
N = 38 25 5 3 10
control
Hemi (ERT)
Het (ERT)
Hemi
Het
C
(LC) CTH C24:1 (1134.9/264.4)
12000
10000
8000
6000
4000
2000
0
N = 38 25 5 3 10
control
Hemi (ERT)
Het (ERT)
Hemi
Het
D
Total CTH
100000
80000
60000
40000
20000
0
N = 38 25 5 3 10
control
Hemi (ERT)
Het (ERT)
Hemi
Het

Figure 23

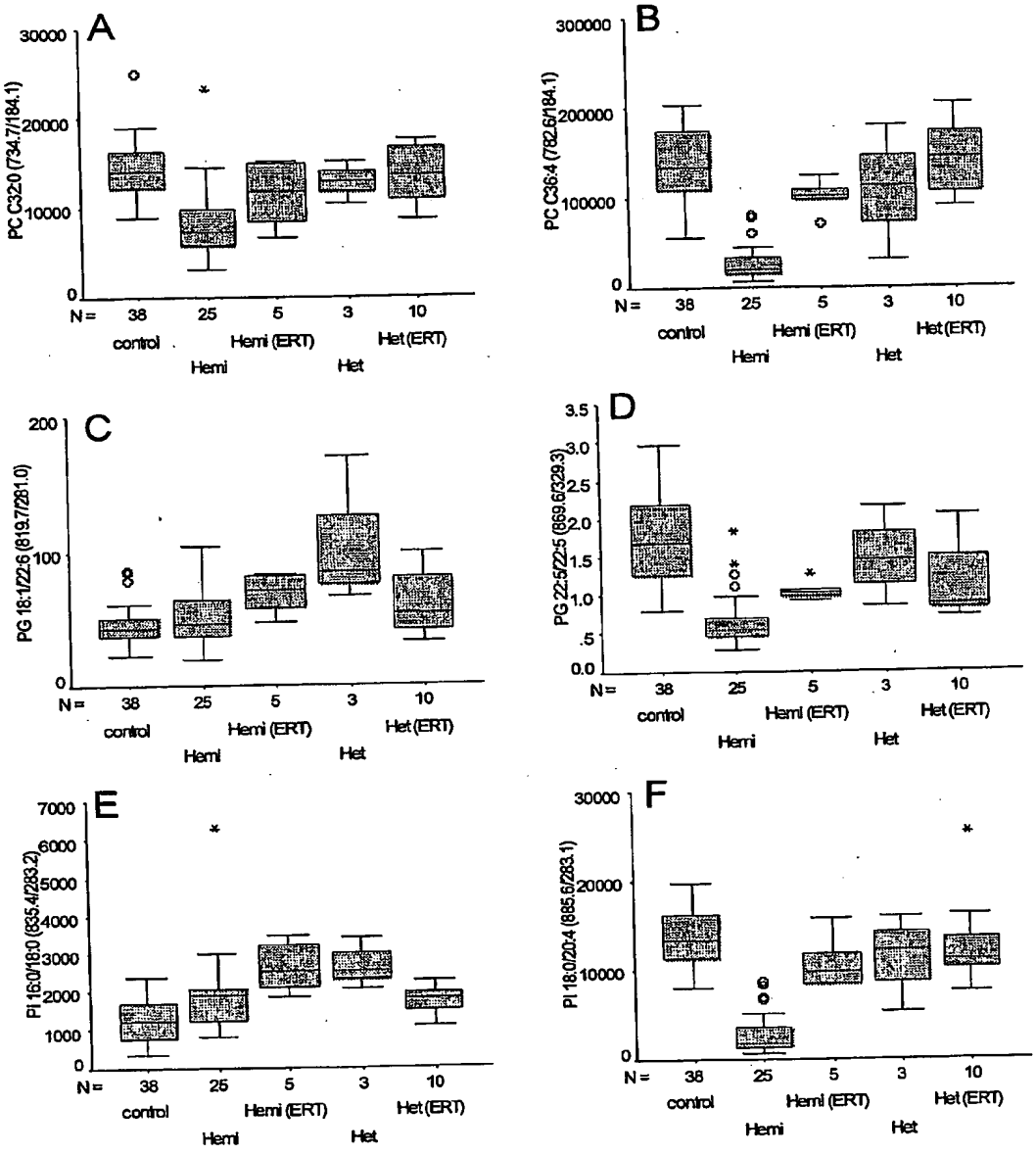
A
PC C32:0 (734.7/184.1)
30000
20000
10000
0
N= 38 25 5 3 10
control Hemi (ERT) Het (ERT)
Hemi Het
B
PC C36:4 (782.6/184.1)
300000
200000
100000
0
N= 38 25 5 3 10
control Hemi (ERT) Het (ERT)
Hemi Het
C
PG 18:1/22:6 (819.7/281.0)
200
100
0
N= 38 25 5 3 10
control Hemi (ERT) Het (ERT)
Hemi Het
D
PG 22:5/22:5 (859.6/329.3)
3.5
3.0
2.5
2.0
1.5
1.0
.5
0.0
N= 38 25 5 3 10
control Hemi (ERT) Het (ERT)
Hemi Het
E
PI 16:0/18:0 (835.4/283.2)
7000
6000
5000
4000
3000
2000
1000
0
N= 38 25 5 3 10
control Hemi (ERT) Het (ERT)
Hemi Het
F
PI 18:0/20:4 (885.6/283.1)
30000
20000
10000
0
N= 38 25 5 3 10
control Hemi (ERT) Het (ERT)
Hemi Het

Figure 24

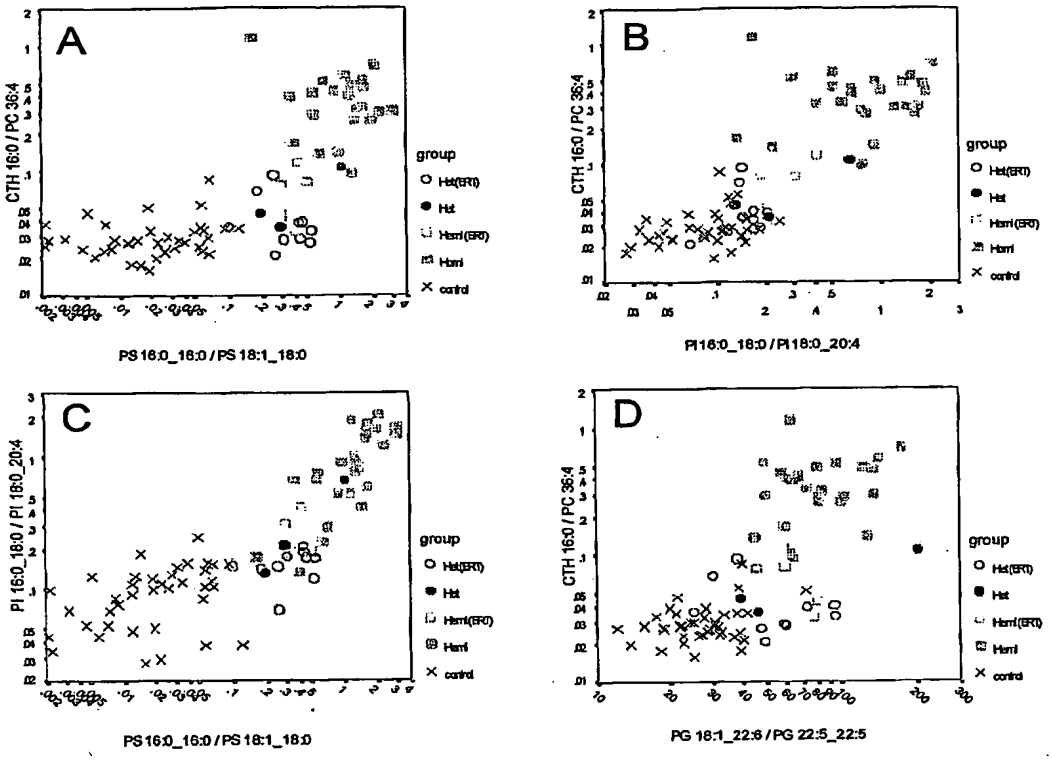
A
CTH 16:0 / PC 36:4
PS 16:0_16:0 / PS 18:1_18:0
group
Ht(ERI)
Ht
Hom(ERI)
Hom
control
B
CTH 16:0 / PC 36:4
PI16:0_18:0 / PI18:0_20:4
group
Ht(ERI)
Ht
Hom(ERI)
Hom
control
C
PI 16:0_18:0 / PI 18:0_20:4
PS 16:0_16:0 / PS 18:1_18:0
group
Ht(ERI)
Ht
Hom(ERI)
Hom
control
D
CTH 16:0 / PC 36:4
PG 18:1_22:6 / PG 22:5_22:5
group
Ht(ERI)
Ht
Hom(ERI)
Hom
control

Figure 25

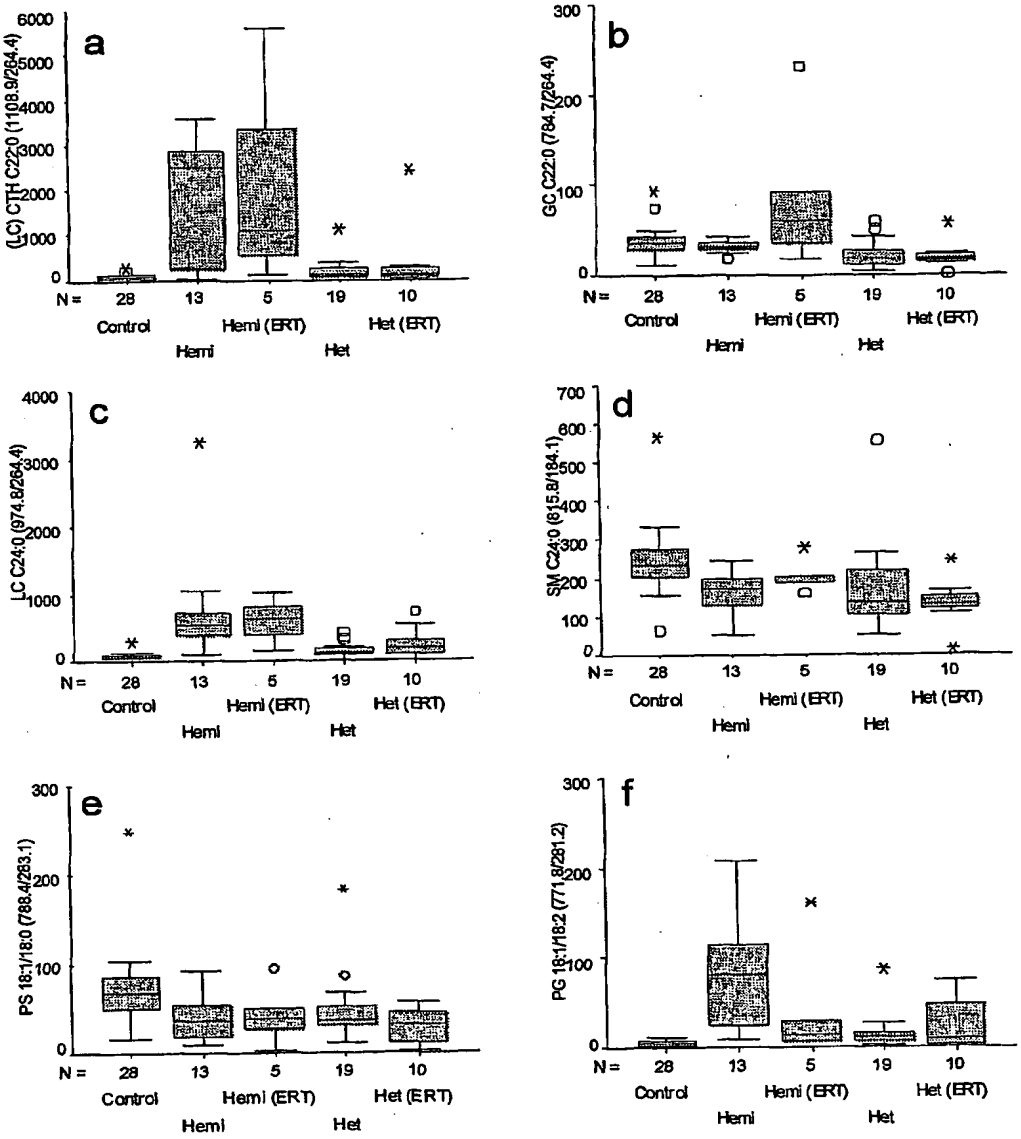
a
(LC) CTH C22:0 (1108.9/264.4)
6000
5000
4000
3000
2000
1000
0
N = 28 13 5 19 10
Control
Hemi
Hemi (ERT)
Het
Het (ERT)
b
GC C22:0 (784.7/264.4)
300
200
100
0
c
LC C24:0 (974.8/264.4)
4000
3000
2000
1000
d
SM C24:0 (815.8/184.1)
700
600
500
400
300
200
100
e
PS 18:1/18:0 (788.4/283.1)
f
PG 18:1/18:2 (771.8/281.2)

Figure 26

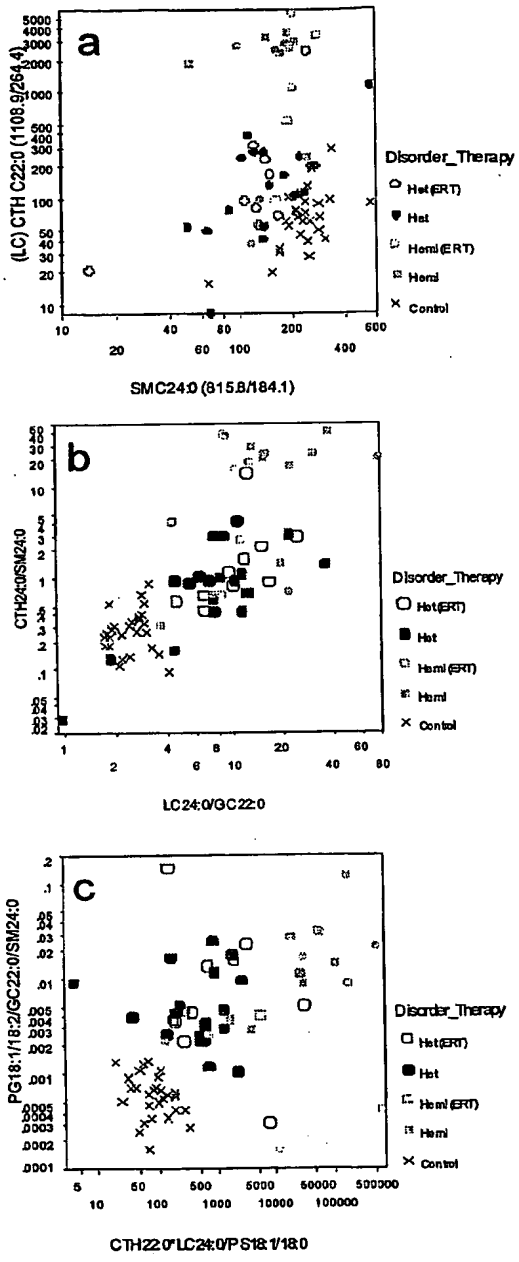
a
(LC) CTH C22:0 (1108.9/264.4)
SMC24:0 (815.8/184.1)
Disorder_Therapy
Het(ERT)
Het
Heml(ERT)
Heml
Control
b
CTH24:0/SM24:0
LC24:0/GC22:0
Disorder_Therapy
Het(ERT)
Het
Heml(ERT)
Heml
Control
c
PG18:1/18:2/GC22:0/SM24:0
CTH22:0*LC24:0/PS18:1/18:0
Disorder_Therapy
Het(ERT)
Het
Heml(ERT)
Heml
Control

Figure 27